



Research Paper

Anesthetic Management Of A Patient With Churg - Strauss Syndrome Who Underwent Laparoscopic Cholecystectomy - Case Report

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ABSTRACT

Churg-strauss syndrome is a rare disease that causes inflammation of small to medium vessels. Diagnostic is mainly clinical with; asthma, eosinophilia and additional vasculitis of multiple organ systems. The anesthetic management of patient with CSS could be complicated by cholinesterase deficiency, hypersensitivity of the airway, chronic steroid usage and multiple organ dysfunctions. Here; we describe our management of a patient with CSS who underwent laparoscopic cholecystectomy under general anesthesia.

The patient had been suffering from asthma for the last 20 years ,peripheral neuropathy and unstable angina and was diagnosed with CSS 2 years previously. During the preoperative period he was on inhaled formoterol, oral prednisone ,dual antiplatelets ,diltiazem ,enalapril were maintained until the operation day, except for enalapril and clopidogrel .In the morning of surgery he was premedicated with midazolam and hydrocortisone. The anesthesia was

induced with propofol, fentanyl and inhaled Isoflurane, any muscle relaxant was not administered during surgery . The surgery was completed uneventfully.

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I. INTRODUCTION

Churg strauss syndrome also known as eosinophilic granulomatosis, was described originally in 1951 (1); as a rare disease that causes inflammation of small to medium sized vessels, associated with antineutrophil cytoplasmic antibodies (ANCA). CSS can affect almost any organ system, the commonest site involved is the lung, others organs involved include skin, peripheral and central nervous system, cardio-vascular system, renal system and gastro-intestinal tract (2, 3). The exact etiology of CSS is unknown, but immune complexes are probably involved (3). Making diagnostic is difficult because patient with CSS have various clinical symptoms and levels of disease progress; the diagnostic is most commonly based on a combination of specific clinical manifestations that distinguish the syndrome from other primary vasculitis (American College of Rheumatology 1990 criteria) (4). Treatment of patient with CSS is directed toward both immediately reducing the inflammation and suppressing the immune system (cyclophosphamide) (5) Patient with CSS display cholinesterase deficiency, airway hyper-reactivity and multi-organ failure (6, 7); for this reasons their anesthetic management could be a real challenge. In the current report we present the anesthetic management of a patient with CSS undergoing laparoscopic cholecystectomy under general anesthesia

II. Case Report:

53 year old male patient, with past history of asthma for the last 20 years , he was currently on inhaled formoterol , the last asthma attack was 3months back. He had history of left lung empyema and bilateral pleural effusion 3 year back, (left more than right) .He was diagnosed with CSS 2 year back when he began to complain of numbness and burning pain of left lowerlimb was evaluated and found to have length dependent sensorimotor axonopathy of Lowerlimb>Upperlimb. He had symptoms such as numbness of left toes progressing upto knee with difficulty in wearing chappals. Nerve biopsy was suggestive of vasculitic neuropathy with peripheral eosinophilia and anti-neutrophil cytoplasmic anti-body (ANCA) was negative. A diagnosis of CSS was then made. He had taken 9 doses of cyclophosphamide intravenously and prednisone 40 mg /day initially, then progressive reduction of the dose; currently 5mg daily. One year back he had an episode of unstable angina, ECG showed

LAHB, ST elevation in V1 and V2, he was heparinised and started aspirin clopidogrel, ACE inhibitor like enalapril and calcium channel blocker -diltiazem. The preoperative physical evaluation showed height and weight of 163 cm and 68kg, respectively. Laboratory studies showed; hemoglobin 12.8 g/dl, total white blood count (WBC) 7800/mm with 500/mm eosinophils, Renal function tests along with serum electrolytes were within normal limits.

ECG showed normal sinus rhythm, T inversion in lead I, II, III aVF, V5, V6

Echocardiography showed EF :40% RWMA of basal, mid inf wall, inferior septum akinetic, basal mid ant wall, anterolateral wall mildly hypokinetic and moderate LV dysfunction, mild MR.

CXR showed Left costophrenic angle blunting and features suggestive of old empyema change. Spirometry showed mild obstructive pattern, Inhaled formeterol and oral prednisone were maintained until the operation day. On the day of surgery, the patient was premedicated with iv midazolam 1 mg and hydrocortisone 100 mg. In the operating room; Routine non-invasive monitoring was established, including non-invasive blood pressure (NIBP), heart rate (HR), pulse oximetry (SpO₂), and electrocardiography (ECG). His baseline blood pressure (BP) and heart rate were 120/75 mmHg and 80/min, respectively. His SpO₂ was 99% while breathing room air and ECG results showed normal sinus rhythm. After obtaining a venous access bolus dose of dexmedetomidine 1mcg/kg was given in 1gm paracetamol infusion 15 min prior to induction. He was preoxygenated with 100% oxygen for 3 min; general anaesthesia was induced with propofol (120 mg) and fentanyl (100 µg); after manual ventilation with oxygen-isoflurane 1% for 3 minutes an endotracheal tube (size 8.5) was placed. Left radial artery cannulated and IBP was monitored. Anaesthesia was maintained on oxygen, N₂O and propofol infusion along with combined infusion of dexmedetomidine 0.5mcg/kg/hr and lignocaine 1.5 mg/kg/hr. Any muscle relaxant was not administered. The surgery was completed uneventfully as ventilation and blood pressure were adequate throughout the whole operation, no complications occurred and total anaesthesia time was 1h and 10 minutes, and any anticholinesterase was administered. At the end of surgery infusions were stopped and tracheal tube was taken out in a deep plane of anaesthesia. Oxygen was supplemented by face mask in post anaesthesia care unit till patient regained full consciousness.

III. Discussion

CSS was first described in 1951 by Dr. Jacob Churg and Dr. Lotte Strauss as a syndrome consisting of asthma, eosinophilia and additional vasculitis of multiple organ systems [1]. The clinical manifestations of CSS usually have three stages [7]. The first phase is called the "allergic" phase and this is characterized by allergic inflammation of the nose, the skin and the lung. People are often diagnosed with late onset asthma during this phase. The second phase is called the "hypereosinophilic" phase, which means that there are too many eosinophils in the body. This phase is characterized by inflammation of the esophagus, stomach or intestine. The third phase is the "systemic vasculitis" phase. During this phase there is inflammation and damage of the blood vessels; the blood vessels in different parts of the body can be damaged. During this phase, the patients with CSS may suffer from fever, weight loss and a lack of energy. These 3 phases are not necessarily contiguous. The American College of Rheumatology (ACR) has offered criteria that must be fulfilled in order to classify a patient as having CSS [8]. In order to be classified as a CSS patient, a patient should have at least 4 of the 6 ACR criteria for CSS to be diagnosed with a sensitivity of 85% and a specificity of 99.7%. These criteria are 1) asthma, 2) eosinophilia [>10% on the differential WBC count], 3) mononeuropathy, 4) transient pulmonary infiltrates on chest X-rays, 5) paranasal sinus abnormalities and 6) a biopsy containing a blood vessel with extravascular eosinophils. Our patient's asthma, peripheral blood eosinophilia, the skin biopsy with an infiltration of eosinophils and the chronic sinusitis fulfilled the ACR diagnostic criteria.

CSS commonly responds to prednisone. High doses of oral prednisone are initially used in an attempt to get the disease into remission as quickly as possible (e.g., oral prednisone 40-60 mg/day). After the first month or so, this high dose of prednisone is slowly tapered down over the ensuing months [9]. Other immunosuppressive drugs such as azathioprine, cellcept, cyclophosphamide or methotrexate may be used in addition to prednisone. High doses of intravenous steroids, and commonly methylprednisolone, maybe useful for those patients with severe disease or for those who are unresponsive to oral prednisone combined with other immunosuppressive medications.

The French Vasculitis Study Group has identified five prognostic factors, the so-called five-factor score (FFS), for patients with necrotising vasculitis, including CSS [10]. These are (1) elevated serum creatinine levels (>1.58 mg/dl), (2) proteinuria (>1 g per day), (3) gastrointestinal tract involvement, (4) cardiomyopathy and (5) central nervous system involvement. The presence of one or more factors is correlated with a 5-year mortality rate ranging from 25.9-46%, whereas an FFS of 0 is correlated with a 5-year mortality rate of 11.9% [10]. The prognosis of patients with CSS is not clearly known, but a 1 year survival rate of 90% and 5 year survival rate of 62% have been reported after treatment with corticosteroid. A significant reduction in mortality has been reported after treatment [11].

Most patients with CSS have asthma. So, pulmonary function tests are essential as part of the preoperative evaluation of CSS patients to assess the risk of perioperative and postoperative pulmonary complications. The patient in this case had been experiencing asthma for the previous 20 years, but his respiratory symptoms were not severe and they were well-controlled with bronchodilators (formeterol 1 puff twice a day), and the pulmonary function tests were within the normal limits.

Surgery requires that a patient have nothing to eat or drink for about 6 hours before anesthesia, with an exception being medication. Patient with CSS and who have been taking corticosteroid for a long time should take their usual medication up until the time of surgery. Adrenal insufficiency could develop, and CSS patients may require perioperative supplemental corticosteroids [12]. The patient in this case took oral prednisolone 10 mg on the morning of surgery and there was no adverse event during the perioperative period. So, we did not medicate him with additional corticosteroid.

For our case, the anesthesia management during the perioperative period was chosen to minimize tracheal activity as the patient had asthma. Propofol reduces the intubation-induced broncho constrictive response [13]. Propofol leads to bronchodilation in patients with chronic obstructive pulmonary disease. The volatile anesthetic agents are bronchodilators that relax the airway smooth muscle and decrease the resistance of the respiratory system. Some opioids can release histamine, and so they can cause bronchospasm. But fentanyl and its analogues have an antihistaminergic action and these may be more effective than morphine for patients with asthma. Furthermore, Groeben reported that the suppression of the cough reflex and the deepening of anesthesia level that are achieved after opioid administration can be helpful in asthmatic patients [14]. The anesthetic management was successful in this case by using propofol, remifentanyl and sevoflurane to induce and maintain anesthesia without respiratory complication.

Taylor and colleagues [15] reported on two patients with CSS who were found to have decreased cholinesterase activity after abnormal sensitivity to suxamethonium was suspected. So, we did not use muscle relaxant as there was no need for muscle relaxation for surgery and our patient was adequately anesthetized using a volatile anesthetic agent and opioid. A deep state of anesthesia was induced and maintained with total intravenous anesthesia. This also reduced any bronchial hyper-reactivity, yet hypotension can occur because of deep anesthesia, so a continuous arterial pressure monitor was used to detect hypotension early. The use of an anticholinesterase agent to reverse muscle relaxation can cause bradycardia, increased secretion and bronchial hyper-reactivity, so we did not use an anticholinesterase agent.

In conclusion, this case was successfully managed using general anesthesia following careful preoperative evaluation and precise perioperative management. The patients with CSS may require intensive care during the course of their disease. Muscle relaxant should be used with caution during surgery, and successful anesthetic management can be achieved.

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